periodic PLEDs, which are often associated with contralateral myoclonic jerks.[2]

The two cases described here illustrate that a diagnosis of Creutzfeldt-Jakob disease should be considered where a rapid decrease in consciousness is accompanied by EEG changes apparently compatible with complex partial status. When there is a clinical suspicion of Creutzfeldt-Jakob disease, the ideal method of monitoring such patients is with continuous EEG recording, allowing documentation of rapid fluctuations. The present cases are atypical in that the progression from presentation to death was rapid, but they underline the fact that minute to minute changes in EEG rhythm, asymmetry, and electrographic responsiveness to benzodiazepines can all be seen in Creutzfeldt-Jakob disease.

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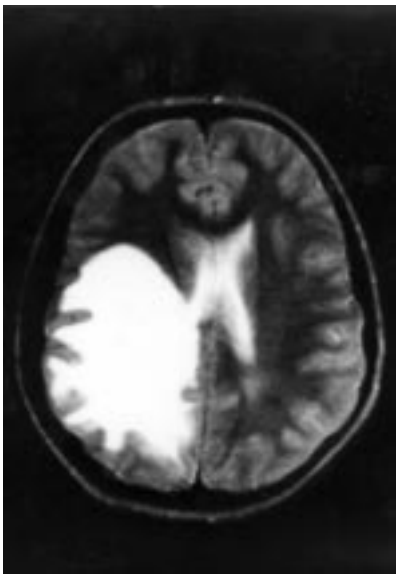
Childhood demyelinating diseases with a prolonged remitting course and their relation to Schilder's disease: report of two cases

Schilder's disease or myelinoclastic diffuse sclerosis is a rare acute or subacute demyelinating disorder which primarily affects children and young adults.[1 2] We report the clinical and neuroradiological follow up of two boys affected by a demyelinating disease with a prolonged relapsing-remitting course, response to corticosteroids, and relatively good long term prognosis.

The first patient presented at the age of 12 with a 2 month history of repeated episodes of headache and blurred vision followed by weakness in the left leg, lasting a few hours. Head CT and bilateral carotid angiography were normal. Two weeks later the left hemiparesis and headache recurred. T2 weighted images on brain MRI disclosed a hyperintense signal in the right parieto-occipital white matter of the centrum semiovale, without a mass effect. Flash visual evoked stimuli elicited a decreased potential on the left side. Motor and sensory nerve conduction were normal. Corticosteroid treatment (prednisone (1mg/kg/day)) reversed the clinical symptoms. At the age of 14, the boy began to have daily headaches. Brain MRI showed the previous white matter lesion, now extending to the parietal, temporal, and occipital areas, with a mass effect and contrast enhancement (figure). Light microscopy of a right parietal stereotactic biopsy disclosed perivascular cuffing with inflammatory cells and ultrastructural examination confirmed a loss of myelinated fibres. Immunohistochemical findings were negative. Five months later the patient had a third episode of left hemiparesis, this time with left sided focal motor and secondarily generalised seizures. Treatment with carbamazepine and cyclophosphamide improved the hemiparesis and controlled the seizures. Brain MRI showed that the mass effect had disappeared, leaving a right parietal, temporal, and occipital lesion. Follow up at the age of 19 showed that, except for a left visual field deficit, the patient had a normal neurological and mental status. He is now receiving azathioprine (75 mg/day).



Brain MRI of patient 1 at the age of 14.5 years. The T 2 weighted image (TR 2000/TE 50) shows a high intensive signal in the parieto-occipital white matter, involving the right centrum semiovale, with mass effect.

The second patient was admitted at the age of 4 because of the sudden onset of headache and vomiting with ataxia and drowsiness followed by generalised clonic seizures. Clinical examination on admission showed left hemiparesis, anisocoria (left>right), and dysarthria. Ocular funduscopy was normal. Head CT disclosed a reduced right lateral ventricle and subarachnoid spaces and, 1 week later, a small hypodense area in the right periventricular white matter. A carotid angiogram was normal. At the age of 5 the child had a second episode characterised by high fever, vomiting, sixth nerve bilateral paresis, dysarthria, truncal ataxia, and stupor. Treatment with corticotrophin (35 units daily for 5 days, then every 48 hours for 20 days) induced a rapid clinical improvement. From the ages of 5 to 14 the child had yearly relapses characterised by the sudden onset of left hemiparesis with variable involvement of the cranial nerves and impairment of consciousness associated with inconsistent alteration of white matter on brain CT (widespread hypodensity in the right centrum semiovale with a mass effect on the right ventricle). These attacks regressed spontaneously or after corticosteroid treatment. The last episode, at the age of 14, consisted of right sided paraesthesias of the face and hand, right hemiparesis, dysarthria, and drowsiness. T2 weighted sequences on MRI disclosed multiple focal abnormalities in supratentorial white matter. Corticosteroid treatment induced marked clinical improvement. Follow up at the age of 18 detected only a bilateral paretic nystagmus and hypometric saccades. Mental development was normal. Brain MRI showed that the white matter lesions had partly regressed.

In both patients the following investigations during and between attacks yielded normal findings: CSF examination (absence of oligoclonal bands), CSF lactate and pyruvate; extensive serological and CSF immune screen; extensive viral, bacterial, and parasitic serological and CSF tests; blood lactate and pyruvate, blood ammonia, amino acids in plasma, urine and CSF, gas chromatography-mass spectrometer analysis of organic acid in urine, adrenal function, plasma C26/C22 fatty acid ratio, serum copper, plasma ceruloplasmin, pyruvate dehydrogenase complex and cytochrome *c* oxidase activity in skin fibroblasts; arylsulphatase A, β-galactosidase, β-glucosidase, and galactocerebroside-β-galactosidase activities in leucocytes.

In both cases the overall findings raise the question of myelinoclastic diffuse sclerosis.[1 2] A prerequisite for the diagnosis is a normal very long chain fatty acid plasma concentration. Clinical signs include an intracranial hypertension syndrome, mental deterioration, hemiplegia, and visual field defects. The disease has either a monophasic course, rarely rapid and fatal, or a relapsing-remitting course.[3] Most patients have neurological sequelae during follow up and few patients fully recover.[3 4] Histological studies typically show a demyelinating process similar to that of multiple sclerosis, with an inflammatory perivascular infiltrate, and in severe cases, cystic lesions. Neuroimaging findings tend to parallel the clinical course. Corticosteroids may improve the outcome of the single relapse and possibly of the disease, as they did in our patients. Some patients respond to immunosuppressive therapy.

In both the patients described the association of headache, signs of diffuse and focal brain dysfunction, a relapsing course, and the response to corticosteroids also raise the possibility of an isolated CNS angiitis, a condition primarily affecting middle aged and elderly people. But neither cerebral angiography nor histological examination disclosed a primary vascular disorder. In addition, the early onset and the sporadic occurrence of the disorder rule out another recently described vasculopathy often associated with familial hemiplegic migraine.[5]

In conclusion, although demyelinating diseases that do not fulfil the classic definition of multiple sclerosis or encephalomyelitis remain difficult to label in children, the two cases we report here seem to fit Schilder's description of myelinoclastic diffuse sclerosis. Owing to the current lack of knowledge on the causes of this disease strict diagnostic criteria cannot be applied. Some presentations may warrant brain biopsy. The differing clinical and neuroimaging features seen in these patients may help in delineating Schilder's disease subtypes.

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Guillain-Barré syndrome after heat stroke

Heat stroke is usually not listed among the events triggering Guillain-Barré syndrome. Two cases of a Guillain-Barré syndrome-like polyneuropathy after heat stroke are on record, although without reference to electroneurography.[1] We report on a patient, who developed Guillain-Barré syndrome 10 days after severe heat stroke. He had electrophysiological evidence of demyelination, increased CSF protein, and high anti-GM1 antibodies. Heat stroke activates the immune system by cytokine release,[2] opens the blood-nerve barrier, and exposes peripheral nerve antigens and thus may induce Guillain-Barré syndrome, as suggested by results from our patient.

A 28 year old drug addict was using anticholinergic drugs against sweating during levomethadone withdrawal. He was found unresponsive in a public garden on a hot summer day (ambient temperature 32°C) after ingesting cocaine. His core temperature was 42.5°C at admission. He was in deep coma with wide unreactive pupils and without corneal and pharyngeal reflexes. Tachypnoea had induced hypocapnia. Blood pressure was 85/30 mm Hg and heart rate was 165/min. He developed disseminated intravascular coagulation, thrombopenia below 10 000 MRD/ml, and metabolic acidosis. Creatine kinase rose from 128 to 751 U/l. Leucocytes and C reactive protein remained normal. After 4 days of coma he was transferred to a closed psychiatry ward because of agitation and frightening hallucinations. After 5 more days he complained of fatigue, myalgia, and arthralgia and, 3 days later, developed fever (38.9°C), tetraplegia, and dysphagia. He required intensive care within hours. Vital capacity was 1.5 l. Proximal arm muscles had MRC grades between 2 and 3. All other limb muscles had grades 1 or 2. Facial muscle weakness increased for 2 more days. There was no ophthalmoplegia, but areflexia and stocking glove hypaesthesia for vibratory and cold stimuli. He did not respond to early intravenous immunoglobulin treatment and underwent plasma exchange from day 14 to 18. Bulbar muscles improved on day 15. Head movements improved 1 week later. Minimal hand functions recurred after 4 more weeks. He still required help with dressing and was unable to stand 14 months after disease onset.

Protein comcentration in CSF was 480 mg/l 2 days after onset of tetraparesis, and 7300 mg/l 2 weeks later. Cell count was 6 cells/mm^3. He had high IgM (500 U/l; enzyme linked immunosorbent assay (ELISA); normal below 120 U/l) but only moderately increased IgA antibodies against GM1. 0nly one of 20 patients with Guillain-Barré syndrome examined in the same laboratory had higher anti-GM1 IgM antibodies. The anti-GM1 antibodies were normal 8 weeks after plasmapheresis.

Compound motor action potentials were<0.7 mV in all tested nerves from day 2 to day 95. Distal latencies were more than 150% above the upper limit of normal in the left peroneal nerve. Conduction velocities were below 70% of the lower limit of normal in the left peroneal and the left median nerve. F latency was above 150% of the upper limit of normal in the left ulnar nerve. F responses were missing in both median nerves and in the right peroneal nerve. A conduction block was present along the right ulnar nerve (wrist stimulation 0.69 mV; plexus stimulation 0.37 mV). Abnormal temporal dispersion and possible conduction block was present in the left ulnar nerve (wrist stimulation amplitude 0.36 mV; duration 8.6 ms; elbow stimulation amplitude 0.19 mV; duration 10.2 ms). Median sensory nerve conduction was normal and sural nerve conduction was moderately slowed (36 m/s) at day 2. Needle EMG disclosed abundant fibrillations and positive sharp waves in proximal and distal limb muscles at day 95.

Decreased sweating due to anticholinergic medication, cocaine induced increased heat production, and high ambient temperature precipitated heat stroke in our patient. Ten days afterwards he developed an acute neuropathy that met clinical and neurophysiological criteria for Guillain-Barré syndrome. Similar time delays have been seen in two other patients with Guillain-Barré syndrome-like neuropathies after heat stroke[1] and in the second of two patients reported as critical illness neuropathy after extreme hyperpyrexia.[3] This patient had increased CSF protein and fasciculations which are unusual in critical illness neuropathy. He may have had Guillain-Barré syndrome as well. Weakness evolved with delay in these four patients with Guillain-Barré syndrome-like neuropathies, whereas it was present immediately after hyperpyrexia in five more patients, who probably did not have Guillain-Barré syndrome. One patient with heat stroke was tetraparetic when he regained consciousness.[4] He had pyramidal and cerebellar signs and persistent atrophic weakness due to axonal or motor neuron loss and no neurophysiological evidence for demyelination. Four of 14 patients with cancer exposed to whole body hyperthermia and chemotherapy complained of weakness immediately after hyperthermia.[5] Their nerve conduction abnormalities are reported as "compatible with scattered demyelination".

Our patient had chronic HCV infection which may be associated with vasculitic neuropathy and cryoglobulinaemia, both absent in our patient. A connection between Guillain-Barré syndrome and non-A non-B hepatitis has been suggested,[6] but the close temporal relation makes heat stroke a more probable cause of the disease in our patient. His high anti-GM1 antibodies suggested immune mediation. Anti-GM1 IgA is increased after *Campylobacter jejuni* infection, whereas IgM dominated in our patient who had no evidence of *Campylobacter jejuni* infection. Heat stroke disrupts the gastrointestinal mucosal wall. Endotoxins enter circulation and stimulate macrophages, which release TNF-α, Il-1, Il-6, and IFN-γ. All these cytokines are raised after heat stroke[2] and open the blood-nerve barrier. This may have exposed the GM1 epitope in our patient. IFN-γ induces Schwann cells to express MHC class II gene product, inviting T cell attack. TNF-α is proinflammatory, myelinotoxic, and increased in Guillain-Barré syndrome.

Guillain-Barré syndrome-like neuropathies have been reported from Saudia Arabia,[1] where heat stroke is common, but they were not noted in connection with epidemic heat stroke in North America.[7] Our patient had all features associated with fatal heat stroke: long lasting coma, shock requiring intravenous catecholamines, metabolic acidosis, and disseminated intravascular coagulation.[7] Guillain-Barré syndrome may occur more often after heat stroke, if more patients survive extreme hyperthermia thanks to intensive care.

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Hydrodynamic performance of a new siphon preventing device: the SiphonGuard

Around 10% to 30% of shunt revisions may be attributed to posture related overdrainage. Of the various siphon preventing devices available at present, two construction types are the most prominent: those using a gravitational mechanism and those using a subcutaneous membrane. Gravitational devices such as Elekta-Cordis Horizontal-Vertical Valve, Chhabra Valve, Fuji Valve, or Miethke Dual-Switch Valve are widely used.[1] Their main drawback is susceptibility to malfunction when the shunt becomes displaced from its vertical axis after implantation and unpredictable operation during persistent bodily movements. The membrane devices: the Anti-Siphon Device (ASD, Heyer Schulte) or Siphon Control Device (SCD, Medtronik PS Medical) have generally proved clinically effective,[2 3] although in some cases these devices may obstruct the CSF drainage when the subcutaneous pressure increases or the scar tissue isolates the device from atmospheric pressure. The flow regulat-